
ANNALS OF THE NEW YORK ACADEMY OF SCIENCES

Volume 460

December 30, 1985

BIOLOGY, CHEMISTRY, AND PATHOLOGY OF COLLAGEN*

Editors and Conference Organizers

RAUL FLEISCHMAJER, BJORN REINO OLSEN, and KLAUS KÜHN

CONTENTS

Preface. By RAUL FLEISCHMAJER, BJORN REINO OLSEN, and KLAUS KÜHN	xiii
---	------

Part I. Collagen Structure

The Structure of Fibril-forming Collagens. By EDWARD J. MILLER...	1
The Structure of Type IV Collagen. By KLAUS KÜHN, ROBERT W. GLANVILLE, WILFRIED BABEL, RUI-QING QIAN, HANS DIER- INGER, TILMAN VOSS, BERNHARD SIEBOLD, ILSE OBERBÄUMER, ULLA SCHWARZ, and YOSHIHIKO YAMADA.....	14
Structure and Macromolecular Organization of Type VI Collagen. By JÜRGEN ENGEL, HEINZ FURTHMAYR, ERICH ODERMATT, HELGA VON DER MARK, MONIQUE AUMAILLEY, RAUL FLEISCHMAJER, and RUPERT TIMPL	25
The Structure of Type IX Collagen. By RICHARD MAYNE, MICHEL VAN DER REST, YOSHIFUMI NINOMIYA, and BJORN R. OLSEN..	38
The Structure of Type VII Collagen. By ROBERT E. BURGESSON, NICHOLAS P. MORRIS, LOUANN W. MURRAY, KEITH G. DUNCAN, DOUGLAS R. KEENE, and LYNN Y. SAKAI.....	47
Structure and Biology of the Globular Domain of Basement Membrane Type IV Collagen. By RUPERT TIMPL, ILSE OBERBÄUMER, HELGA VON DER MARK, WOLFRAM BODE, GEORG WICK, SABINE WEBER, and JÜRGEN ENGEL	58
Supramolecular Collagen Assemblies. By BARBARA BRODSKY and ERIC EIKENBERRY	73

*The papers in this volume were presented at the Conference on Biology, Chemistry, and Pathology of Collagen, which was held by the New York Academy of Sciences on October 17-19, 1984 in New York City.

Part II. Collagen Genes, Structure and Regulation

The Structure of the Chicken $\alpha 2$ Collagen Gene. By HELGA BOEDTKER, MITCHELL FINER, and SIRPA AHO	85
Isolation and Characterization of the Human Fibrillar Collagen Genes. By FRANCESCO RAMIREZ, MICHAEL BERNARD, MON-LI CHU, LEON DICKSON, FRANK SANGIORGI, DOMINIQUE WEIL, WOUTER DE WET, CLAUDINE JUNIEN, and MARK SOBEL.....	117
Structure of the Type II Collagen Gene. By WILLIAM B. UPHOLT, CHARLES M. STROM, and LINDA J. SANDELL	130
Short-Chain Collagen Genes and Their Expression in Cartilage. By BJORN R. OLSEN, YOSHIFUMI NINOMIYA, GUILLERMINA LOZANO, HIROSHI KONOMI, MARION GORDON, GRAHAM GREEN, JANEY PARSONS, JEROME SEYER, HILLARY THOMPSON, and GEORGE VASIOS.....	141
Structural and Functional Analysis of the Genes for $\alpha 2(I)$ and $\alpha 1(III)$ Collagens. By BENOIT DE CROMBRUGGHE, AZRIEL SCHMIDT, GENE LIAU, CHIAKI SETOYAMA, MARIA MUDRYJ, YOSHIHIKO YAMADA, and CATHERINE MCKEON.....	154
Structure and Expression of the Collagen Genes of <i>C. Elegans</i> . By DAVID HIRSH, GEORGE N. COX, JAMES M. KRAMER, DAN STINCHCOMB, and RICHARD JEFFERSON.....	163

Part III. Collagen Biosynthesis

Regulation of Collagen Biosynthesis: Heparin Alters the Biosynthetic Phenotype of Vascular Smooth Muscle Cells. By RICHARD A. MAJACK and PAUL BORNSTEIN.....	172
Biosynthesis and Properties of Procollagens V. By JOHN H. FESSLER, NANCY SHIGAKI, and LISELOTTE I. FESSLER.....	181
Post-Translational Processing of Procollagens. By KARI I. KIVIRIKKO and RAILI MYLLYLÄ.....	187
Collagen Synthesis in Virus-transformed Cells. By SHERRILL L. ADAMS, MAURIZIO PACIFICI, RICHARD J. FOCHT, EILEEN S. ALLEBACH, and DAVID BOETTIGER	202
Anchorin CII, a Type II Collagen-binding Glycoprotein from Chondrocyte Membranes. By KLAUS VON DER MARK, JÜRGEN MOLLENHAUER, PETER K. MÜLLER, and MICHAEL PFÄFFLE	214
The Role of Lymphokines and Monokines in Fibrosis. By SHARON M. WAHL.....	224

Part IV. Developmental Biology of Collagen

Are Collagen Fibrils in the Developing Avian Cornea Composed of Two Different Collagen Types? Evidence from Monoclonal Antibody Studies. By THOMAS F. LINSSENMEYER, JOHN M. FITCH, JEROME GROSS, and RICHARD MAYNE	232
--	-----

Collagen Fibrillogenesis in Human Skin. <i>By</i> RAUL FLEISCHMAJER, JEROME S. PERLISH, and RUPERT TIMPL	246
Fibroblasts Create Compartments in the Extracellular Space Where Collagen Polymerizes into Fibrils and Fibrils Associate into Bundles. <i>By</i> DAVID E. BIRK and ROBERT L. TRELSTAD	258
Tissue-specific Gene Expression in Mouse F9 Embryonal Carcinoma Cells: Type IV Collagen. <i>By</i> MARKKU KURKINEN, DENISE P. BARLOW, and BRIGID L. M. HOGAN	267
Endothelial Cell-Extracellular Matrix Interactions. <i>By</i> BRUCE M. PRATT, DAVID FORM, and JOSEPH A. MADRI	274

Part V. Genetic and Acquired Collagen Diseases

Mutations in Osteogenesis Imperfecta Leading to the Synthesis of Abnormal Type I Procollagens. <i>By</i> DARWIN J. PROCKOP, MON-LI CHU, WOUTER DE WET, JEANNE C. MYERS, TAINA PIHLAJANIEMI, FRANCESCO RAMIREZ, and MARIA SIPPOLA	289
Molecular Basis of Clinical Heterogeneity in the Ehlers-Danlos Syndrome. <i>By</i> PETER H. BYERS and KAREN A. HOLBROOK	298
Collagenase in Recessive Dystrophic Epidermolysis Bullosa. <i>By</i> EUGENE A. BAUER	311
Biochemical and Immunohistochemical Studies on the Scirrhous Carcinoma of Human Stomach. <i>By</i> Y. NAGAI, H. SUNADA, J. SANO, S. ONODERA, K. ARAI, H. KONOMI, T. MINAMOTO, R. HATA, H. HORI, I. NAKANISHI, H. KITAOKA, and G. SAKAMOTO	321
Role of the Extracellular Matrix in Cancer. <i>By</i> LANCE A. LIOTTA and C. N. RAO	333
Collagenase in Rheumatoid Arthritis. <i>By</i> CARLO L. MAINARDI	345
Type II Collagen-induced Arthritis. <i>By</i> JOHN M. STUART, ALEXANDER S. TOWNES, and ANDREW H. KANG	355
The Involvement of Type IV Collagen in Goodpasture's Syndrome. <i>By</i> JÖRGEN WIESLANDER and DICK HEINEGÅRD	363
Collagen Synthesis by Scleroderma Fibroblasts. <i>By</i> THOMAS KRIEG, JEROME S. PERLISH, CORNELIA MAUCH, and RAUL FLEISCHMAJER	375

Poster Papers

Biosynthesis and Structure of Collagen VI Produced in Human Skin Fibroblast Cultures. <i>By</i> MONIQUE AUMAILLEY, HELGA VON DER MARK, and RUPERT TIMPL	387
Collagen Degradation in Human Lung Fibroblasts Is Time Dependent. <i>By</i> FRANK A. BARILE and ROBERT S. BIENKOWSKI	390
Biosynthesis of a Disulfide-bonded Type X Collagen in Bovine Growth Plate Cartilage. <i>By</i> R. I. BASHEY, M. REMINGTON, C. T. BRIGHTON, and S. A. JIMENEZ	392

Intracellular Degradation of Newly Synthesized Collagen. <i>By</i> RICHARD A. BERG and DONALD S. NEBLOCK.....	395
Type IX Collagen Identified As Proteoglycan Lt. <i>By</i> PETER BRUCKNER, LLOYD VAUGHAN, and KASPAR H. WINTERHALTER.....	397
Synthesis of Type X Collagen in Developing Chick Embryo Tibiae. <i>By</i> R. CANCEDDA, O. CAPASSO, P. CASTAGNOLA, N. QUARTO, F. DESCALZI-CANCEDDA, G. PONTARELLI, and E. GIONTI.....	399
Binding of Laminin to Type IV Collagen: A Morphological Study. <i>By</i> ARISTIDIS S. CHARONIS, EFFIE C. TSILIBARY, PETER D. YURCHENCO, and HEINZ FURTHMAYR.....	401
Partial Purification and Characterization of Precursors to Minor Cartilage Collagens. <i>By</i> CHARLES C. CLARK and CHARLOTTE F. RICHARDS.....	404
Glucocorticoid Regulation of Procollagen Synthesis. <i>By</i> KENNETH R. CUTRONEO, KENNETH M. STERLING, JR., SUSAN SHULL, DEBRA COCKAYNE, and SHARON ILLEYNE.....	407
Sequence Analysis of Chicken Type II Procollagen cDNA Clones Containing Helical-Domain Coding Sequences. <i>By</i> FERENC DEÁK, W. SCOTT ARGRAVES, IBOLYA KISS, KENNETH J. SPARKS, and PAUL F. GOETINCK.....	409
Morphometric Analysis of Defective Collagen Fibril Growth in Bovine Osteogenesis Imperfecta (Australia Type). <i>By</i> L. J. DENHOLM, C. E. HALL, R. R. MINOR, and L. P. KROOK.....	412
Electronmicroscopic Localization of Deletions in the Human Pro- α 2(I) Collagen Gene. <i>By</i> WOUTER DE WET, MARIA SIPPOLA, MICHAEL BERNARD, DARWIN PROCKOP, MON-LI CHU, and FRANCESCO RAMIREZ.....	415
Measurement of Collagen Deposition Following a Standardized Subcutaneous Injury. <i>By</i> ROBERT F. DIEGELMANN, WILLIAM J. LINDBLAD, and I. KELMAN COHEN.....	419
Isolation and Characterization of Mammalian Parent Type M (IX) Collagen. <i>By</i> VICTOR C. DUANCE, SANDRA F. WOTTON, and ALLEN J. BAILEY.....	422
Mechanical Properties of Connective Tissues: Analysis of Genetic Disorders. <i>By</i> MICHAEL G. DUNN, SATISH KALATH, WEI-CHORNG LAI, PETROS TSIPOURAS, FREDERICK H. SILVER, and NOSHIR LANGRANA.....	426
Type VI Collagen: Studies on Its Localization, Structure, and Biosynthetic Form with Monoclonal Antibodies. <i>By</i> EVA ENGVALL and HELENA HESSLE.....	429
Physiological and Biochemical Characterization of Avian Dystrophic Muscle Reveals Alterations of Collagen. <i>By</i> HOWARD FEIT and MASATAKA KAWAI.....	431
Sponge Collagens: Structural Diversity and Fibril Assembly. <i>By</i> ROBERT GARRONE, PASCAL GUÉNIN, and MARLÈNE MAZZORANA.....	434
Abnormal Accumulation of Basement Membrane (Type IV) and Cytokeletal (Type V) Collagens in the Strictures of Crohn's Disease: The Probable Role of Intestinal Smooth Muscle Cells. <i>By</i> M. F.	

GRAHAM, R. F. DIEGELMANN, C. O. ELSON, S. GAY, and R. GAY.....	439
Partial Characterization of Collagen Types IX and X Synthesized by Embryonic Chick Chondrocytes. By M. E. GRANT, C. M. KIELTY, A. P. L. KWAN, D. F. HOLMES, and S. L. SCHOR.....	443
Defective Prolyl-4-Hydroxylase Activity Associated with Defective Differentiation in Response to Retinoic Acid in a Mutant F9 Mouse Teratocarcinoma Stem Cell Line. By LORRAINE J. GUDAS, MICHAEL ROGUSKA, and SHO-YA WANG.....	445
Identification of Novel Human Basement Membrane Zone Components by Monoclonal Antibodies. By D. W. HOLLISTER, L. Y. SAKAI, and R. E. BURGESSON.....	449
Transcriptional Control of Human Diploid Fibroblast Collagen Synthesis by γ -Interferon. By SERGIO A. JIMENEZ and JOEL ROSENBLOOM.....	453
An Electron Microscope Study of the First-Formed Aggregates in Collagen Fibril Assembly <i>in Vitro</i> . By K. E. KADLER and J. A. CHAPMAN.....	456
Normal and Mutant Human Collagen Genes. By R. KAUFMAN, E. HENKE, M. DUVIC, F. BISAT, and S. PINNELL.....	461
Basement Membrane Supramolecular Complexes. By HYNDA K. KLEINMAN, GORDON W. LAURIE, JOHN R. HASSELL, FRANCES B. CANNON, and VICKI L. STAR.....	463
Characterization of a Connective Tissue Glycoprotein Which Inhibits Collagen Synthesis by Scleroderma Fibroblasts. By F. X. MAQUART, J. CORNILLET, G. BELLON, A. RANDOUX, J. P. BOREL, and B. KALIS.....	466
Animal Models with Heritable Defects in the Formation and/or Degradation of Collagen Fibrils. By RONALD R. MINOR, JOYCE A. M. WOOTTON, and DONALD F. PATTERSON.....	469
Role of the Basal Lamina Collagen (Type IV) in Brain Vascular Permeability. By M. MISKULIN, G. GODEAU, J. M. TIXIER, L. ROBERT, and A. M. ROBERT.....	471
Treatment of Chondrocytes with β -Glycerophosphate Increases Synthesis of Short-Chain (Type X) Collagen. By JENNY B. MORRIS and GARY BALIAN.....	475
Annelid Cuticle Collagen: A Very Long Protein. By LOUANN W. MURRAY, JÜRGEN ENGEL, and MARVIN L. TANZER.....	478
A Homozygous Frameshift Mutation in the Pro- α 2(I) Collagen COOH-Propeptide Results in Osteogenesis Imperfecta. By JEANNE C. MYERS, LEON A. DICKSON, F. MICHAEL POPE, VEIKKO R. KORHONEN, ALAN NICHOLLS, DARWIN J. PROCKOP, and TAINA PIHLAJANIEMI.....	482
Monoclonal Antibodies to Collagen Hydroxylases and Their Use for the Localization of the Genes for These Enzymes on Human Chromosomes. By RAILI MYLLYLÄ, LEILA PAJUNEN, MATTI HÖYHTYÄ, TAINA TURPEENNIEMI-HUJANEN, KARL TRYGGVASON, and KARI I. KIVIRIKKO.....	486

Expression of Two Unusual Collagens in Chemically Transformed Syrian Hamster Cells. <i>By</i> B. PETERKOFKY, G. MAJMUDAR, J. BATEMAN, and W. PRATHER	489
Serum Collagenase Activity in Pregnant, Parturient, and Postpartum Women. <i>By</i> MOHAMMAD RAJABI, DAVID D. DEAN, and J. FREDERICK WOESSNER, JR.	492
Changes in the Nuclear and Cytoplasmic Levels of Type I and Type II Collagen RNAs during Growth of Chondrocytes in 5-Bromo-2'-Deoxyuridine. <i>By</i> STEPHEN A. SAXE, LEWIS N. LUKENS, and PHILIP J. PAWLOWSKI	494
Type X Collagen within the Embryonic Chick Sternum: Immunohistochemical Localization with a Monoclonal Antibody. <i>By</i> THOMAS M. SCHMID, EILEEN GIBNEY, and THOMAS F. LINSSENMEYER...	497
Dietary Ascorbic Acid Increases Nascent Collagen Production in Skins of Diabetic Rats by Reversing Procollagen Underhydroxylation and Concomitant Intracellular Degradation. <i>By</i> MICHAEL SCHNEIR, LORNE GOLUB, and NANGAVARAM RAMAMURTHY...	500
Covalent Structure of Collagen: Salient Differences in the $\alpha 2(I)$ Polypeptide Chain. <i>By</i> JEROME M. SEYER and ANDREW H. KANG ..	503
A Structurally Abnormal $\alpha 2(I)$ Collagen Chain in a Further Patient with the Ehlers-Danlos Syndrome Type VII. <i>By</i> BEAT STEINMANN, VELIDI H. RAO, and RICHARD GITZELMANN.....	506
Cloning of Type III Collagen and Expression in Aortic Smooth Muscle Cells. <i>By</i> MARY ANN STEPP, MARK KINDY, CARL FRANZBLAU, and GAIL SONENSHEIN.....	510
Altered Type I/Type III Procollagen mRNA in Cultured Keloid Fibroblasts: Evidence for Regulation on the Transcriptional Level. <i>By</i> JOUNI UUITTO, ANDREA J. PEREJDA, R. PATRICK ABERGEL, MON-LI CHU, and FRANCESCO RAMIREZ.....	514
Human Placental $\alpha 1(V)\alpha 2(V)\alpha 3(V)$ and $[\alpha 1(V)]_2\alpha 2(V)$ Collagen Heterotrimers. <i>By</i> MICHEL VAN DER REST, CHRISTOPHER NIYIBIZI, and PETER P. FIETZEK.....	517
Studies on the Distribution of Hydroxypyridinium Cross-Links in Different Collagen Types. <i>By</i> JIANN-JIU WU and DAVID R. EYRE ..	520
Gene Structure: Cartilage and Basement Membrane Collagen Genes. <i>By</i> YOSHIHIKO YAMADA, KIMITOSHI KOHNO, YOSHIKI SAKURAI, PILAR FERNANDEZ, ANNE MARIE NUNEZ, SEISHI KATO, and GEORGE R. MARTIN	524
A 210,000-Dalton Pepsin-resistant Fragment of Type IX Collagen. <i>By</i> NATSUO YASUI, PAUL D. BENYA, and MARCEL E. NIMNI	527
Type IV Collagen "7S" Tetramer Formation: Aspects of Kinetics and Thermodynamics. <i>By</i> PETER D. YURCHENCO and HEINZ FURTHMAYR.....	530
Index of Contributors	535

